Case Report

Anaesthetic management of modified radical mastectomy in a patient with carcinoma breast with Bernard–Soulier syndrome

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Abstract

Bernard-Soulier syndrome (BSS) is a rare congenital bleeding disorder characterised by thrombocytopenia, large platelets and a prolonged bleeding time. Patients with Bernard–Souliersyndrome are at risk of severe bleeding. A mutation involving glycoprotein complex 1b-V-IX renders platelets unable to bind Von Willebrand factor (vWF) and form clots. Patients often report spontaneous epistaxis, bleeding gums or heavy menstrual periods. Bernard–Soulier syndrome patients require rigorous perioperative planning.[1] We present a female patient with BSS that underwent bilateral modified radical mastectomy for her bilateral carcinoma breast. Search words: giant platelet syndrome, Bernard–Soulier syndrome, breast cancer, bleeding diasthesis, peri operative management.

Keywords: radical mastectomy, bernard–soulier syndrome.

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INTRODUCTION

Bernard–Soulier syndrome is a rare congenital bleeding disorder characterised by thrombocytopenia, large platelets and a prolonged bleeding time.[2] The incidence in the USA is 1:1 000 000.[3] However, inherited as an autosomal recessive condition, it is commoner in populations with high incidence of consanguineous marriage[4] The underlying defect in Bernard–Soulier syndrome is the absence or decreased expression of the GPIb/IX/V complex on the surface of the platelets. This complex is the receptor for Von Willebr and factor, this results in deficient binding of VWF to the platelet membrane causing defective platelet adhesion[5] The definitive diagnosis of Bernard–Soulier syndrome is made by identifying isolated defective ristocetin-induced agglutination when placed in an aggregometer. The diagnosis may be confirmed biochemically or by genotyping[3].

CASE REPORT

A 38-year-old woman presented for bilateral modified radical mastectomy for bilateral carcinoma breast .She had been diagnosed with Bernard–Soulier syndrome at the age of 4. Previous hospital record showed history of torrential bleed following tooth extraction at age 9, requiring admission and blood transfusion. She also gave history of having excess menstrual bleed for which she has been on oral contraceptives since the age of 15. She was nulliparous. She had received three cycles of chemotherapy every 21 days consisting of 5 fluorouracil, epirubicin, cyclophosphomide. She was hypertensive, on amlodipine 10 mg and atenolol 50 mg twice daily. Preoperative consultation was done with haematologist for work up and optimisation. Preoperative coagulation and haematological investigations were as follows: Hb 11 g/dl, platelet count was 35 t x10^9/L on automated counts and was 90x10^9/L with giant platelets on manual counting, PT 9 seconds (control 11.2), APTT 27 seconds (control 28.5). The smear identified anisocytosis, Howel Jolly bodies, burr cells, schistocytes, basophilic stippling and poikiolocytosis. Manual count of platelets was done on the day of surgery 92x10^9/L.Pre operative work up

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included ECG, ECHO, chest Xray, renal function test and liver function test which were all unremarkable. Two large bore ivcannulae were used for venous access. Patient received tranexamic acid (25 mg/kg) and desmopressin(0.3 µg/kg).The patient was anaesthetised with 150 mg propofol and 120 µg fentanyl, and maintained with isoflurane 1.4% in 50% oxygen. Tracheal intubation was facilitated with vecuronium 6mg. Controlled hypotension was maintained using nitroglycerine infusion tirated to maintain mean arterial pressure 55-60 mm Hg. Meticulous use of cautery was done by surgeon to minimise blood loss. Judicious use of crystalloids were used to avoid dilutional thrombocytopenia. During the 4-hour surgery, 2 units of packed red blood cells, 8 units of platelets were transfused guided by TEG. Following surgery, two drains were left in situ, which drained a total of 800 ml over a period of four days. During this time, the patient was transfused 3 units of packed red blood cells, and 8 units of platelets. Hb was 10 gm%, platelets count of 80 $x10^9$/L on manual count. The drains were removed on day 3 and the patient was discharged on day 5.

DISCUSSION

A constant feature is the presence of a small number of very large platelets with a rounded shape (main volume 11–16 **µ**m^3; diameter 4–10 **µ**m). Platelet counts typically range from 20,000 to 100,000/**µ**l. The very large platelets in Bernard–Soulier syndrome which are often mistaken for lymphocytes in automatic counters and may show platelet counts lower than actual. manual counting is essential for an accurate determination of platelet count in these patients. In Bernard–Soulier syndrome, symptoms are those of frequent nosebleeds, bleeding from gums, and easy bruising. Bleeding often starts in childhood. Torrential bleeds are associated with major trauma or surgery. Women usually suffer with menorrhagia and severe bleeding may occur after childbirth. Patients may be first diagnosed in pregnancy[6] The early use of recombinant factor VIIa at doses of 90 to 100 µg/kg as a first-line therapy, alongside platelet transfusion may be used in an attempt to reduce the perioperative use of blood products.[7] The limiting factor for the use of recombinant factor VIIa is the cost factor, with each microgram costing nearly 1$,[8] Use of high doses like 90 to 100 µg/kg would mean exorbitant hospital expenditure which is out of reach for patients in developing countries belonging to low socio economic status. Transfusions of allogeneic platelets remain the definitive treatment for patients with BSS.[9] By doing manual count of platelets on the day of surgery unnecessary pre operative platelet transfusion may be avoided which are based on automated count. Desmopressin DDAVP nasal sprays prior to sugery was used along with tranexamic acid in addition to controlled hypotension to minimise intra operative bleeding. DDAVP use in bleeding disorders is well reported[10] DDAVP binds V2 receptors found on endothelial cells to increase vWF release and promote Factor VIII.[11] Tranexamic acid (TA) is an anti-fribinolytic that inhibits the activation of plasminogen to plasmin[12]

CONCLUSION

This is the report of a successful bilateral modified radical mastectomy on an adult having Bernard-Soulier syndrome with bilateral carcinoma breast. Pre-operative planning is imperative with consultation between Anaesthesiologist, haematologist and surgeon. The utility of anti-fibrinolytics and DDAVP in BSS patients is clearly a cost effective way of managing these patient posted for surgery.

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